CASE REPORT

Reticular Dystrophy of the Retinal Pigment Epithelium

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A laser flare-cell meter was used to examine blood–aqueous barrier function in a 57-year-old male patient who had typical reticular dystrophy of the retinal pigment epithelium with a bilateral hyperpigmented reticular pattern at the posterior pole in both eyes. Fluorescein angiography showed hypofluorescent reticular net resembling a fishing net with knots associated with a central diffuse hyperfluorescence. Dark adaptation test revealed that the final rod threshold was elevated. Laser photometry showed an increase in the aqueous flare intensity in both eyes, as compared to the results in normal subjects. Quantitative measurement of the aqueous flare intensity by a laser flare-cell meter might indicate abnormalities in the blood–aqueous barrier in patients with reticular dystrophy of the retinal pigment epithelium. [*J Formos Med Assoc* 2007;106(6):490–494]

Key Words: blood–aqueous barrier, reticular dystrophy of the retinal pigment epithelium

In 1950, Sjögren[1] first described "dystrophia reticularis laminae pigmentosae retinae" in eight of 13 children from a Swedish family. The fundus lesions were characterized by a reticular network of black hyperpigmented lines covering most of the posterior pole. Pigmented knots present at the intersection of the dark lines caused the fundus to resemble a fishing net with its knots.

Retinitis pigmentosa has been shown to cause dysfunction of the blood–aqueous barrier.[2] In this study, we evaluated the function of the blood–aqueous barrier in a patient with reticular dystrophy of the retinal pigment epithelium to determine whether this disease has the same trends as retinitis pigmentosa.

Laser photometry now provides a noninvasive technique to assess blood–aqueous barrier function.[3,4] In this report, we describe the use of a laser flare-cell meter to examine abnormalities in the blood–aqueous barrier in this rare disease.

Case Report

A 57-year-old man was referred to us for evaluation of unusual lesion of the fundus in August 2003. There was no family history of night blindness or consanguity. His past medical history was unremarkable.

Examination showed a visual acuity of 20/20 in both eyes. Biomicroscopy showed a clear cornea and media in both eyes. An ophthalmoscopic examination showed a bilateral symmetrical hyperpigmented reticular pattern at the posterior pole extending beyond the vascular arcade and nasally to the optic disk. Hyperpigmented knots were noticed at the junction where hyperpigmented lines

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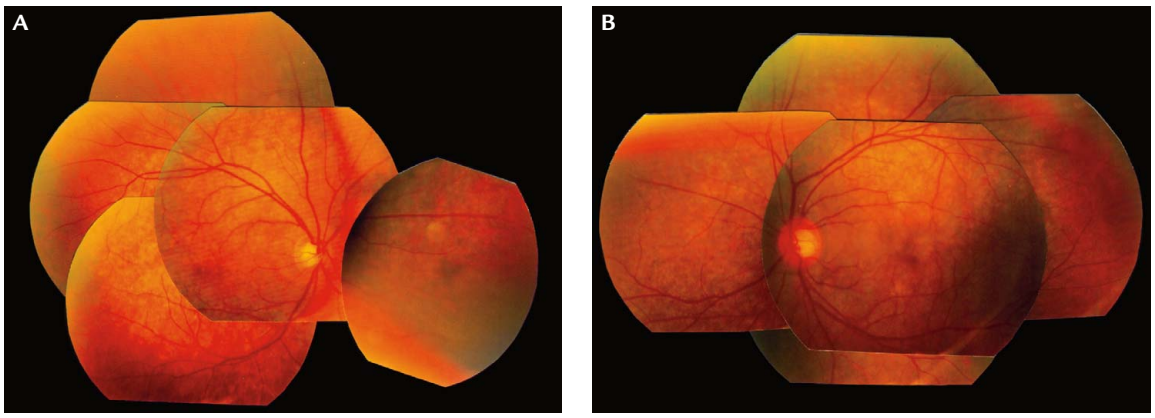

Figure 1. Reticular pattern of pigment epithelial hyperpigmentation morphologically like a fishing net with knots: (A) right eye; (B) left eye.

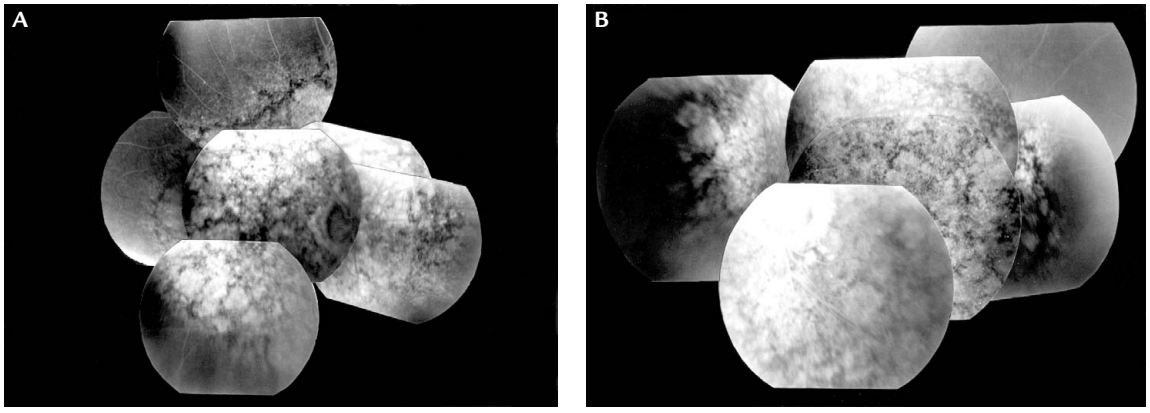

Figure 2. Fluorescein angiography shows hypofluorescent reticular net resembling the meshes of a fishnet associated with a central diffuse hyperfluorescence: (A) right eye; (B) left eye.

crossed. Alternate areas of pigment epithelial hyperpigmentation and hypopigmentation formed a reticular pattern analogous to the meshes of a fishnet. The optic disks and retinal vessels were normal (Figure 1). Fluorescein angiography showed bilateral symmetrical hypofluorescent reticular net with an evident dark spot at the junctions, resembling a fishing net with knots, corresponding to zones of hyperpigmentation. A central diffuse hyperfluorescence corresponding to zones of hypopigmentation was also noticed. There was no leakage of fluorescein at any time (Figure 2).

Examination of color vision with the Farnsworth-Munsell 100-Hue test revealed normal results in the right eye with a total error score of 68 and abnormal results in the left eye with a total error score of 116 and a tritian orientation to the errors (Figure 3). Dark adaptation studies with the Goldmann-Weeker adaptometer revealed the final rod threshold was elevated 2.2 log units in the right eye and 2.4 log units in the left eye (Figure 4). Visual fields performed on the Goldmann perimeter revealed normal peripheral fields without central scotomas. Electroretinographic responses were normal in both eyes. The electrooculogram was normal with light peak/dark trough values of 2.05 in the right eye and 2.07 in the left eye.

The aqueous flare intensity was measured with a laser flare-cell meter (FC 1000; Kowa, Tokyo, Japan) 30 minutes after pupillary dilatation with 0.5% tropicamide and 5% phenylephrine hydrochloride. There was a marked increase in the aqueous flare values in both eyes; 15.7 ± 3.2 (mean $\pm$ standard deviation) photoncounts/msec in the right eye and 12.8 ± 3.7 photoncounts/msec in the left eye. In 10 age-matched control subjects,

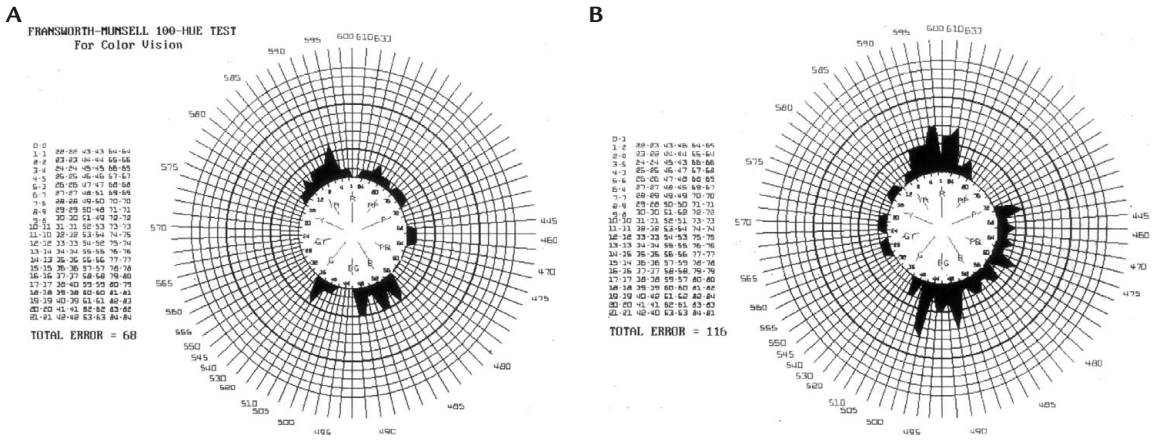

Figure 3. Farnsworth-Munsell 100-Hue test: (A) total error score of 68 in the right eye; (B) total error score of 116 with a tritian orientation in the left eye.

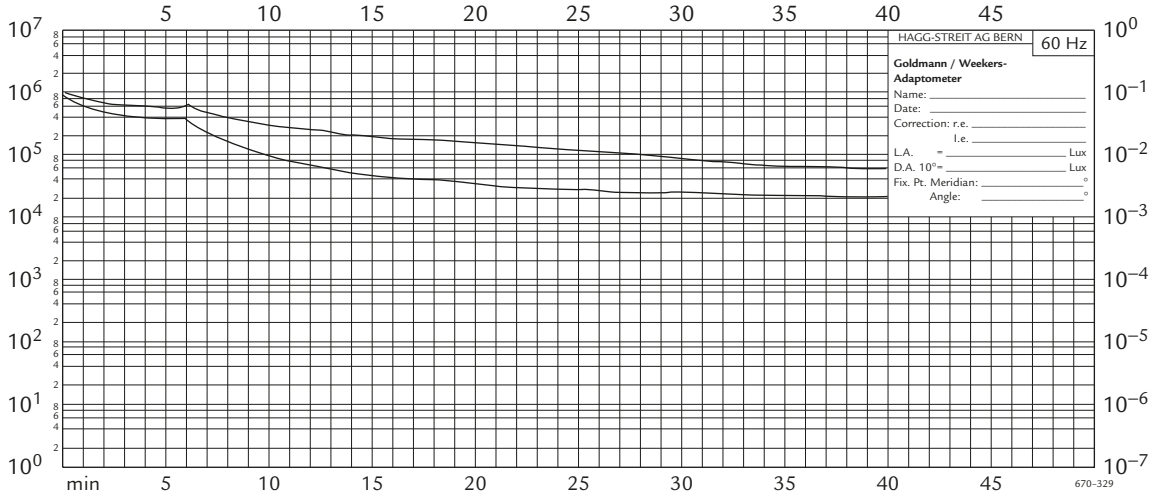

Figure 4. Dark adaptation test shows elevated final rod threshold of 2.2 log units in the right eye (lower curve) and 2.4 log units in the left eye (upper curve).

the aqueous flare intensity averaged 4.60 ± 1.04 photoncounts/msec.

Discussion

Sjögren's original report demonstrated that reticular dystrophy of the retinal pigment epithelium has a progressive course. In the initial stage, an accumulation of pigment is present in the fovea. From there, it migrates in the direction of the foveal periphery and a reticular pigmented network, resembling a fishnet with knots. In the advanced stage, the network extends and covers an oval area of most of the posterior pole. In the more advanced stage, the network becomes bleached and irregular and small white dots appear in the retinal pigment epithelium.[1]

Fluorescein angiography demonstrates a mosaic pattern, showing hypofluorescent bands or segments surrounded by areas of nonleaking hyperfluorescence. This would indicate reticular bands of pigment epithelial hyperpigmentation surrounded by halos of hypopigmentation. The choroidal vessels appear to be normal. The ophthalmoscopic and fluorescein angiographic findings in our patient were identified to those in other reports.[5–8]

According to the retinal function tests in patients with reticular dystrophy of retinal pigment

epithelium, the color vision is usually normal but might also be abnormal. The color vision function in this patient was normal in the right eye and mildly abnormal in the left eye. Dark adaption test in patients with reticular dystrophy of retinal pigment epithelium has been reported as normal or subnormal. This patient had an elevated final rod threshold indicating abnormal dark adaptation, which might be a parameter of this disease. Electroretinography is usually normal but might be abnormal in this disease. This patient had a normal electroretinography. Electrooculogram might be normal or abnormal in this disease. This patient had a normal electrooculogram.

Reticular dystrophy of the retinal pigment epithelium has been shown to be associated with spherophakia with myopia and luxated lenses, partial atrophy of the iris, scleral staphyloma, convergent strabismus and choroidal neovascularization.[1,8] General abnormalities reported in conjunction with reticular dystrophy of the retinal pigment epithelium include deaf-mutism and choreatiform behavior.[1]

Three types of ophthalmic dystrophies of the retinal pigment epithelium, including reticular dystrophy, butterfly-shaped dystrophy and macroreticular dystrophy, have been defined as pattern dystrophy of the retinal pigment epithelium.[9,10] Retinal function tests of these three types are essentially normal, except for the electrooculogram in butterfly-shaped dystrophy. These three types of pattern dystrophy have been reported to be present in affected families, suggesting that they may represent variations in the expressivity of the same dystrophy.[9,10]

Differential diagnosis between reticular dystrophy of retinal pigment epithelium and other types of retinal pigment epithelial dystrophies is necessary. The fundus pictures of the changes in the fovea can be easily distinguished from other hereditary central dystrophies such as Stargardt's disease, vitelliform dystrophy, dominant drusen and dominant cystoid macular edema. Reticular dystrophy of the retinal pigment epithelium has to be differentiated from other types of flecked retina. Fluorescein angiography can demonstrate the typical pictures of reticular dystrophy of the retinal pigment epithelium, including hypofluorescent reticular net resembling a fishing net with knots associated with a central diffuse hyperfluorescence. These pictures can aid differential diagnosis with the other types of flecked retina.

Patients with retinitis pigmentosa have been demonstrated to have dysfunction of the blood–aqueous barrier.[2] However, no studies of the function of the blood–aqueous barrier in reticular dystrophy of the retinal pigment epithelium have been reported. Usually, indirect measurements, inferences from fluorescein angiography, or anterior chamber coefficients of fluorescein leakage values have been used as an index of blood–aqueous barrier permeability.[11] However, fluorophotometric analysis with systemic fluorescein administration is complicated by rapid metabolism of the dye and the adverse effects of fluorescein. With the recent development of laser photometry, noninvasive assessment of the function of the blood–aqueous barrier is now more feasible.[3,4] The laser flare-cell meter consists of a helium–neon laser slit lamp and binocular microscope equipped with a photomultiplier and a personal computer to analyze the data. The instrument allows measurement of the protein concentration in the aqueous humor *in vivo* by measuring light scattered from aqueous protein.

The aqueous flare intensity in this patient was elevated about 3.4 times in the right eye and about 2.8 times in the left eye, compared with normal values obtained from age-matched control subjects. The increase in the aqueous flare intensity in this patient shows that the function of the blood–aqueous barrier may be altered in reticular dystrophy of the retinal pigment epithelium. The sites of the breakdown of the blood–aqueous barrier remain unknown, but are possibly at the stroma of the iris root and in the iris vessels.[12] The changes may be mediated by chemical mediators,[13–15] and may also be influenced by neurogenic or humoral factors.[16] Meanwhile, the proteins released by the breakdown of the blood–retinal barrier may diffuse anteriorly into the anterior chamber directly, or these released factors

may have a direct effect on the vascular permeability of the anterior segment. In this patient, fluorescein angiography demonstrated hypofluorescent reticular net with a central diffuse hyperfluorescence indicating the possible presence of the breakdown of the blood–retinal barrier which might further influence the blood–aqueous barrier function in reticular dystrophy of the retinal pigment epithelium. This needs further clarification in the future.

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